

A Discussion of Meniere's Disease (Endolymphatic Hydrops)

Inner Ear Anatomy and Function

The inner ear is a delicate membraneous sense organ, which is encased in a bony shell. It is suspended within a lattice-like bony framework, called the mastoid bone, which is located behind the outer ear. The delicate inner ear membranes are surrounded by a fluid called perilymph, which is rich in sodium, and lies between the outer bony layer and the inner membraneous layer. Within the membraneous layer circulates a second fluid rich in potassium, called endolymph. The electrical potential created by the separation of these two fluids is the driving force of the inner ear.



The organ of hearing is called the cochlea, which resembles a snail with its 2 1/2 turns; it is divided into three chambers or scala. The scala media is separated from the scala tympani by the basilar membrane and from the scala vestibuli by Reissner's membrane. Only the scala media contains endolymph, whereas the other two contain perilymph. The organ of Corti is housed within the scala media, and rests on the basilar membrane. Special cells with hair-like projections are attached to the tectorial membrane, and project to the hearing nerve (cochlear nerve). The nerve travels through the temporal bone (the bone surrounding the outer, middle and inner ear) to the brainstem. During its course through the temporal bone, it is joined by the two balance nerves (vestibular nerves) and the facial nerve. All four travel together to the brainstem. Upon entering the brainstem, the cochlear nerve relays its message to nerves that will travel to that particular region of the brain responsible for decoding the message (temporal lobe).

The Mechanics of Hearing



Hearing occurs as sound enters the outer ear canal and causes vibration of the tympanic membrane (eardrum) and subsequently the movement of the middle ear bones. The piston-like action of the stapes bone (stirrup) initiates a fluid wave within the perilymph of the scala vestibuli. This "traveling wave" in turn activates the hair cells of the organ of Corti, causing the nerve to discharge. The hair cells are responsible for converting the mechanical energy of the fluid wave into an electrical signal, which will be processed by the brain. There are actually two different types of hair cells in the cochlea: inner and outer hair cells. Inner hair cells are the special sensory receptors that receive the traveling wave information and relay it to the brainstem. The outer hair cells are responsible for amplifying the traveling wave signal, and "fine tuning" the signal to a frequency specific region of the cochlea.

Hearing loss may arise from the outer, middle and inner ear, as well as from the cochlear nerve, brainstem or temporal lobe. A loss that arises as a result of a blockage of sound energy from reaching the inner ear is referred to as conductive; whereas, a loss that results from injury to the inner ear or central structures is called sensori-neural. Although a clinical exam will often identify the type of hearing loss, an audiogram (hearing test) is usually necessary to pinpoint the

location, as well as the severity. Specialized hearing tests, and (uncommonly) imaging studies are sometimes performed if the cause of the loss is still unknown.

The Mechanics of Balance



The balance portion of the inner ear is made up of three semicircular, fluid filled canals which join a larger, globular structure called the vestibule. Similar to the cochlea, the semicircular canals are tubular structures filled with endolymph and surrounded by perilymph. The hardest bone in the body, the labyrinthine bone, surrounds the entire structure. Each semicircular canal is oriented at right angles to the others, comprising vertical, horizontal and posterior (behind) canals. The right and left semicircular canals are mirror images of each other, so that each and every direction of angular head motion is represented by both ears. At the junction of each semicircular canal and vestibule is a special receptor for angular rotational movements of the head, referred to as the crista. The crista contain hair cells embedded in a gelatinous matrix, with accompanying nerve fibers. As the head turns in a particular direction, the fluid within that semicircular canal turns in the opposite direction bending the hair cells and inducing a neural discharge. That signal is sent through the vestibular (balance) nerve to the brain where it is interpreted, and adjustments are made in eye movements and postural control. This ensures that the eyes remain on a given target, and that the arms and legs remain in a good position for maintaining stable posture. Within the vestibule are hair cells that respond to changes in head and body movements in the horizontal and vertical planes. These "otolithic" hair cells are embedded in a layer of calcium carbonate, making them top heavy and therefore motion sensitive for both linear acceleration and gravitational forces.

Nerve fibers from the crista and the otolithic organs form two large balance nerves, the superior and inferior vestibular nerves. They travel from the inner ear to the brainstem along with the cochlear and facial nerves. Within the brainstem they form an extensive neural network involving nerves from the eyes, ears, the cerebellum, and positional receptors "proprioceptors" located in the arms, legs and neck. The brain interprets this information, and makes modifications in eye, head and body position to maintain a fixed eye position, and erect posture. Unfortunately, there are also connections to the thalamic region of the brain, which is responsible for the nausea and vomiting which accompanies most disturbances within the vestibular system. A sensation of dysequilibrium may accompany any imbalance or dysfunction within this neural network. Therefore, similar symptoms of imbalance or "dizziness" may be experienced by injury to the eye, ear, brain, and proprioceptors from the extremities. Therefore it is often difficult to determine the site of injury based on symptoms alone, and diagnostic testing is necessary.



Meniere's Disease

Prosper Meniere was a French physician who described a new disease process in the mid-1800's. The disease he identified consisted of four symptoms: attacks of vertigo, ear fullness or pressure, low-pitched tinnitus (ringing), and fluctuations in hearing. At the time, the cause of the ailment was unknown, but Meniere believed that it originated within the ear. Since that time much has been learned about the disease and its treatment; interestingly however, we still do not know the cause of the disease Meniere described. There are now known to be multiple variants of this disease process, with similar pathologic consequences (abnormal tissue changes). Some forms

affect only hearing, others only balance and still others have a few, but not all of the classic four symptoms.

Histopathology



It has been well characterized since the 1930's, that all forms of Meniere's disease involve dilation of the scala media, with subsequent bulging of Reissner's membrane into the scala vestibuli. Eventually the membrane ruptures with mixing of endolymph and perilymph, causing injury to the hair cells, producing the characteristic symptoms. This process is called "endolymphatic hydrops", meaning too much endolymph. Only the disease process described by Meniere himself should be referred to as Meniere's disease, whereas all the others are more properly termed hydrops, or endolymphatic hydrops. The precise cause of the excessive fluid is unknown, whether due to an overproduction of endolymph or from an under-resorption is hotly debated. Either way, the net result is too much endolymph with distressing symptoms. Although the etiology (cause) of "Meniere's disease" is unknown, there are many known causes of endolymphatic hydrops. These include allergy, immune mediated, metabolic disorders, infections (syphilis), congenital malformations of the ear and trauma. The known causes occur infrequently compared to the idiopathic form (unknown cause), but a search is often undertaken.

Diagnosis

The discussion you have with your physician is the most important aspect of making the diagnosis. The vertigo spells are always episodic, never occurring more than once a day, and often separated by days, weeks or even years. Vertigo (sensation of motion) in hydropic attacks can occasionally be heralded by ear fullness, an increase in ringing (tinnitus) or a sudden drop in hearing. Rarely, an improvement in hearing precedes the attack (Lermoyez syndrome). The attack typically lasts anywhere from 20 minutes to an entire day, depending on how long it takes for Reissner's membrane to repair itself. The vertigo is typically described as a spinning sensation (either the environment or the individual spins), which is quite disabling, and often associated with severe nausea and vomiting, sweating and occasionally loss of bowel and/or bladder control. They are not associated with severe headaches. The Tumarkin variant of hydrops has sudden loss of postural control rather than vertigo, thought to be due to involvement of the otolithic component of the inner ear. After an attack of vertigo, one may experience a period of unsteadiness lasting from hours to weeks (occasionally years) as the brain compensates for the loss of balance function.

Hearing loss in hydrops is quite variable, ranging from a sudden, severe loss without return of function, to a gradual decline over months to years. The most typical pattern is that of hearing fluctuations, with periods of good and poor hearing, associated with a gradual decline in word recognition. The classic form of Meniere's disease consists of a low frequency loss during an attack that may (or may not) return to normal afterwards. Essentially, any pattern of hearing loss may be associated with hydrops; therefore, the audiometric pattern can not be relied upon to make the diagnosis. A demonstrable fluctuation in hearing in one ear or the other helps to make the diagnosis of hydrops.

Classically, low-pitched roaring sound is heard in the affected ear prior to, or during an attack of vertigo. This too is highly variable, and some individuals may experience constant ringing in the ear (tinnitus). The precise etiology of tinnitus is poorly understood, but thought to arise from a loss of the usual spontaneous activity of the cochlear hair cells. A sensation of fullness or

pressure in the ear may accompany the vertigo attack as well. This is thought to arise from over-distension of the endolymphatic compartment with injury to the hair cells.

Not unusually, patients with hydrops are quite sensitive to changes in barometric pressure, and may experience more attacks in the spring and fall. Hormonal changes that occur during menses and pregnancy may trigger an attack. Tension, stress, anxiety, allergy and exhaustion are also known triggers of an attack.

Testing

A long list of disease processes must be excluded prior to making the diagnosis of hydrops or Meniere's disease. Tests of hearing, balance, blood count and chemistry and occasionally imaging, are often performed. Pure tone and speech audiometry, as well as electrocochleography (ECoG), are routinely performed. The former is a standardized hearing test, while the latter is a more specific test of inner ear fluid balance. Balance testing is performed if the diagnosis is in doubt, or persistent dysequilibrium is present. When the diagnosis of hydrops is entertained, an MRI or CT scan (imaging studies) is performed to ensure that the symptoms are not being caused by a tumor. It has been shown that nearly 1 patient in 10 with a tumor on the balance nerve will demonstrate "classic Meniere's symptoms". This does not mean that 1 out of 10 people with hydrops has a tumor, as these tumors are exceptionally rare, yet it remains an integral part of the diagnostic workup.

Treatment

Each and every therapeutic endeavor employed is aimed at eliminated vertigo. Hearing loss, fullness, pressure and tinnitus that occur with hydrops often defy known treatment, although a few options are available. With any given treatment, some patients will have improvement in these symptoms, while others will have no change, and still others may have aggravation of these symptoms. This likely represents the natural history of the disease process. The mainstay of treatment for all forms of hydrops is a low sodium diet and a diuretic. We encourage our patients to maintain a very low sodium diet, less than 1800 mg of sodium each day. A pamphlet will be provided to assist you with this. Additionally, a diuretic is often used to eliminate extra fluid. Both of these maneuvers are employed to help maintain fluid balance within the inner ear. As many as 85% of patients will benefit from this treatment, either with complete cessation of vertigo, or (at least) substantially reduced attack severity. Some individuals are extremely salt sensitive, having a vertigo attack within hours of sodium indiscretion (hamburger and fries for example). Occasionally, a sudden drop in hearing will respond to a steroid burst over 7-10 days. With progression of the disease, this often becomes ineffective. Hundreds of other forms of medical therapy have been tried and shown to be no better than placebo (sugar pill). A vertiginous attack may be controlled through a variety of vestibular suppressants, depending on the age and health status of the individual.

For those patients who do not respond to medical therapy alone (approximately 15%), surgical treatment is necessary to control the vertigo. Four surgical options are available to everyone, although those with good hearing are advised to consider only those options which attempt to preserve normal hearing.

Endolymphatic Shunt Surgery

This minimally invasive procedure has been hotly debated for many years. It involves an incision behind the ear and a mastoidectomy (removing the bone behind the ear). The endolymphatic sac

is then identified within the mastoid bone, which is connected to the inner ear endolymphatic compartment. The sac is opened and a piece of surgical grade plastic is inserted inside. The theoretical purpose of this operation is to "shunt" excessive endolymph away from the inner ear. In reality, no one knows why the operation is so successful. Perhaps it has to do with altering the vascularity or immune function of the sac, but it is doubtful that it has anything to do with "shunting" fluid. It has been shown countless times that this operation is successful in controlling vertigo in 75% of those with hydrops. The surgery has relatively few risks, and can be performed as an outpatient.

Vestibular nerve section

Attacks of vertigo can be eliminated by cutting the balance nerves on the affected side. The operation can be performed either from above the ear, or behind the ear. It requires a craniotomy (opening through the skull) to access the balance nerves as they exit the inner ear. The vestibular nerves are separated from the cochlear and facial nerves, and then divided. Because of the intracranial nature of this operation, the patient is observed in the intensive care unit overnight, and remains in the hospital from 3 to 5 days. The risks of the operation are higher than for an endolymphatic shunt, however there is a much higher rate of vertigo control, up to 97%.

Labyrinthectomy

For an individual with very poor hearing, this is the procedure of choice. The operation begins with a mastoidectomy, but then continues deeper to remove the inner ear semicircular canals and vestibule. Some surgeons also cut the balance nerves at the same time. Vertigo is controlled up to 99% of the time with minimal risks, unfortunately hearing on the side of the surgery is eliminated.

Intratympanic gentamycin

Gentamycin is an antibiotic which has as a side effect, injury to the balance organs. This can be utilized to eliminate vertigo in many individuals with hydrops. The eardrum on the side of the disease is anesthetized (local anesthesia), and a tiny needle is passed into the middle ear space. A small amount of this antibiotic is injected into the middle ear, and the patient remains in a recumbent position for 20-30 minutes. Anywhere from 2-5 injections over the course of a month are often necessary to affect vertigo control. Between injections hearing and balance function are monitored. At the first sign of either hearing loss or balance dysfunction, the treatments are discontinued. Studies have shown that control of vertigo can be accomplished in up to 80% of patients, however hearing loss occurs in as many as 20% as well. For many elderly and medically infirm patients, this is an excellent option.

Prognosis

Hydrops is a chronic disease, like diabetes or hypertension. It needs to be treated medically (low sodium diet and diuretic) like other disease processes for many years. Like other disease states, it can be well controlled with therapy. Certain individuals with hydrops will develop the disease in the opposite ear. This occurs in 4-10% of patients with hydrops. This fact needs to be taken into account when surgical options are entertained.